Treatment Options for Symptomatic Degenerative Joint Disease Secondary to Legg-Calvé-Perthes Disease

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Abstract

The treatment options for neglected Legg-Calvé-Perthes disease (LCPD) leading to symptomatic degenerative joint disease always have posed a challenge for the orthopedic surgeon. In addition, the literature is disorganized in this regard. Therefore, a structured literature review of the treatment options for the symptomatic sequelae of LCPD, especially in the context of joint replacement and resurfacing, is very much needed.

The radiologic features associated with Legg-Calvé-Perthes disease (LCPD) first were described by Henning Waldenstrom in 1909 in Stockholm. Because he attributed the findings to tuberculosis infection he did not receive the credit. In 1910, the condition was described independently by Arthur Legg from the United States, Jacques Calvé from France, and Georg Perthes from Germany.[1]

Pathophysiology

Legg-Calvé-Perthes disease is idiopathic avascular necrosis of the femoral head occurring in childhood. This disease is not a single avascular episode, however, but is the result of sequential infarcts. Though LCPD usually is a self-limiting condition, it often leads to changes in the proximal femoral architecture and acetabulum, which can result in secondary degenerative joint disease (DJD), usually in the third or fourth decade of life.[2] Symptoms may occur at an earlier age in the absence of arthritic change caused by instability of the hip from acetabular dysplasia.

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Etiology

Thrombophilia, acetabular retroversion, mutation in type 2 collagen gene, mechanical weakness of the femoral head, exposure to second-hand smoke, and trauma all have been postulated as possible etiologies.[3-9] However, no conclusive evidence to support these theories has been found. Despite the common endpoint, as with many diseases, the etiology is likely to be multifactorial with different contributions from genetic, constitutional, and environmental factors, which may differ geographically.

Classifications

A number of classification systems have been proposed for LCPD to assess the prognosis and best treatment for different disease patterns. A classification system based on the extent of involvement of the capital femoral epiphysis and head-at-risk signs was introduced by Lloyd-Roberts and colleagues.[10] This classification system was modified by Caterall, but it usually cannot be applied early in the disease process.[11] The Salter and Thompson classification system, based on the extent of subchondral fracture in the superior dome of the femoral head, can be applied early in the disease process to determine the treatment strategy.[12] Unfortunately, the subchondral fracture only is visible on standard anteroposterior and lateral radiographs in about 15% of patients. Sensitivity can be improved by repeat radiographs in multiple planes, but this would increase the exposure to radiation.

Herring's lateral pillar classification, based on the height of lateral pillar of the femoral head, is widely used and is a good predictor of the amount of head flattening at skeletal maturity;[13] this is because deformity, once present, does not seem to remodel significantly, thus loss of height or flattening of the femoral head is a good measure of the final deformity. Use of the Herring system, however, does not allow for treatment to prevent deformity from occurring. The Stulberg classification system, based on congruency between the femoral head and acetabulum once healing is complete, seems to accurately predict the risk of developing degenerative arthritis in the long term.[14]

The Waldenstrom stages classification describes the chronology of the disease process from onset through sclerosis, fragmentation, healing, and, finally, remodeling of the femoral head.[1]

Evaluation

All of the above classification systems require regular biplanar radiographs. Other modalities also may be utilized in the diagnosis, staging, and preoperative planning of the disorder, however.

Bone scanning can be used to obtain a reasonable estimate of the degree of femoral head involvement, but it involves a substantial radiation dose and only shows the situation at a single point in time. Because LCPD involves sequential infarcts, a false conclusion may be made from a single scan. Bone scanning is easier to use in unilateral LCPD because of the availability of the patient's normal hip for comparison. The technique can be helpful in making a diagnosis of avascular bone when symptoms are present but changes are not yet visible on conventional radiographs. Computed tomography with reconstruction may be used to evaluate femoral head shape for preoperative joint replacement planning and the need for special implants. However, it is not very helpful in managing the early stages of LCPD.

Magnetic resonance imaging (MRI) has been tested in LCPD but has been found to be of little value in determining the extent of femoral head involvement because the degree of edema surrounding the infarct makes the damage seem far greater than it actually is.[15,16] Furthermore, similar to a bone scan, a single MRI gives only a snapshot view of a sequential process. Repeat MRI scans may be more helpful. Because it is possible to have small bone infarcts, which do not progress to LCPD but heal, MRI can give false-positive results.

Early Treatment Options for LCPD

The long-term results of LCPD have shown abnormal changes in the proximal femoral anatomy and acetabulum, leading to early development of arthritic changes in the hip.[2] The goal of early treatment of LCPD, therefore, is to prevent femoral and acetabular deformity, which, subsequently, may cause debilitating destruction of the hip joint by a combination of poor containment and movement of the hip. Theoretically, if a softened femoral head can be contained within the spherical acetabulum, it should not deform. The acetabulum is only big enough to cover a third of the femoral head, however. Full and free movement of the proximal femoral epiphysis in the acetabulum allows molding of the whole head and prevents flattening of the epiphysis, which may result in progressive lateral subluxation and a roller-bearing joint with hinge abduction. Even though the femoral head invariably heals with some degree of coxa magna causing relative acetabular dysplasia, a well-contained and mobile joint should remain congruent. Furthermore, there is some potential for acetabular remodeling, which may diminish the degree of dysplasia, though the potential for acetabular remodeling decreases with increasing age. Grzegorzewski and colleagues suggested that even an already-deformed head might have an improved outcome if it is contained in the acetabulum.[17] However, others have noted that deformity, once present, does not improve to a significant extent. Indeed, a recent multicenter evaluation of outcomes, by Herring and colleagues, found no difference in outcome for children diagnosed under age 8 years between those with untreated LCPD and those managed by conservative or surgical containment methods.

The type of treatment for LCPD depends on the child's age, the extent of head involvement, and the stage of the disease at the time of the evaluation. Traditionally, nonoperative management is applicable to children under age 5 or 6 years or with less than half head involvement. Treatment options include bed rest, skin traction, and abduction bracing for a variable period of time. Surgical treatments, traditionally used for children over age 6 years with more than half head involvement, include varus osteotomy with or without derotation, innominate osteotomy, or a shelf procedure. Only the shelf procedure has been shown to significantly alter acetabular remodeling and prevent progressive head deformity. Schneidmueller and colleagues claimed good results for treating overgrowth of the greater trochanter in children and adolescents with congenital hip conditions, including LCPD by trochanteric epiphyseodesis (which only works if done at age 6 years or younger), distal transfer of the greater trochanter, and femoral neck lengthening osteotomies.[18] There is no point in treating LCPD in the healing stage or later since it is too late to alter the outcome. Valgus osteotomy is reserved for cases of hinge abduction.

Joint Arthroplasty Options for LCPD

Arthroplasty procedures, which usually include total hip arthroplasty (THA) or resurfacing hip arthroplasty (RHA), in LCPD patients with advanced symptomatic DJD, frequently are employed but fraught with unique challenges. Apart from developing coxa vara, coxa plana, and a short, broad anteverted femoral neck, these patients are young and may have had previous surgery with retained hardware. Leg-length discrepancy, acetabular dysplasia, and retroversion also may pose serious challenges during the hip surgery.

As a treatment modality, THA can be very effective in addressing the abnormal femoral head and neck in the treatment of the symptomatic sequelae of LCPD. A classification system based on the site, geometry, and etiology of the deformity, put forth by Berry from the Mayo Clinic, is useful in the preoperative planning of the procedure in these patients. The classification system basically divides the site of deformity into greater trochanter, femoral neck, metaphysic, and diaphysis; the geometry of deformity into torsional, angular, translational, and size abnormality; and the etiology of deformity into developmental, metabolic, previous osteotomy, and previous fracture.[19]

An overhanging trochanter and a high-riding trochanter are 2 common problems encountered when dealing with an abnormal proximal anatomy of the femur. An overhanging trochanter may lead to varus migration of the femoral implant and a high-riding trochanter can impinge on the pelvis. These problems can be managed by a transtrochanteric approach and advancement of the trochanteric position at the completion of the total hip replacement.

A varus femoral neck can be managed with offset femoral stems, which may equalize leg length and improve the abductor mechanism. The use of modular femoral components may better accommodate the abnormal proximal femur anatomy. If the acetabulum is deficient, an acetabuloplasty or acetabular augmentation can be combined with THA.

Our literature review revealed a very limited number of articles addressing the role of THA for the treatment of patients with symptomatic DJD secondary to LCPD. Kawasaki and colleageus found that operating time and perioperative blood loss were significantly higher in patients who received THA after a failed transtrochanteric rotational osteotomy for avascular necrosis compared with patients who received THA without previous osteotomies. The patients' Harris hip scores, stability of implants, and survival rates were not significantly different, however.[20]

Wangen and colleagues studied the long-term results of THA with a hydroxyapatite-coated stem in patients younger than age 30 years (their patient group included 3 LCPD patients).[21] They had some mechanical failures at the acetabular side, but, generally, had excellent results with a fully hydroxyapatite-coated femoral stem with no revisions after 10 to 16 years of follow-up.

Resurfacing hip arthroplasty, as a treatment for younger LCPD patients who have developed secondary osteoarthritis, has enjoyed recent resurgence. It is a bone-conserving procedure, and, therefore, very attractive for young patients who may require surgeries in the future. It inherently increases joint stability when compared with THA and later can be converted to total hip replacement, if the need arises.

Boyd and colleagues compared a series of 19 RHAs with the results of standard THA found in the literature and reported comparable results. The RHAs provided equally satisfactory Harris hip scores, range of motion, and leg-length equalization.[22]

A short neck length and low head-to-neck ratio poses a major challenge during the resurfacing procedure. Amstutz and colleagues reported the head-neck ratio, anterior offset, lateral offset, and posterior offset in a cohort of 14 hips with LCPD that underwent surface arthroplasty. They noted a lateral offset ratio averaging 0.12, which may indicate tilting of the femoral head medially. The authors found a significant improvement in the range of flexion and internal rotation after the resurfacing procedure.[23] It was pointed out that if notching could not be avoided, it preferably should be on the medial side where the cortex is thicker and stronger. Avoidance of notching of the neck on the anterior and lateral tension sides can reduce the incidence of complications. The restoration of head-neck ratio and offset usually can be done with thinner acetabular components and the use of femoral components up to the size of 56 mm.

O' Hara recently reported a 1- or 2-stage osteotomy procedure to normalize the neck before undertaking the resurfacing. He proposed a concurrent osteotomy and trochanteric osteotomy with advancement as a 1- or 2-stage procedure before proceeding with the resurfacing.[24]

Since the femoral head-neck unit is abnormal in these patients and the head-neck unit is preserved during the resurfacing procedure, these patients especially are prone to the femoroacetabular impingement and restricted range of motion. This can be avoided by removing any anterior osteophytes or anterior translation of the femoral component.[25]A posterior approach and a trochanteric slide also have been found to be helpful in avoiding impingement.[26]

In contrast to conventional THA, RHA usually is unable to correct significant leg-length discrepancy, is a more technically challenging procedure with a steep learning curve, and is difficult to perform unless the deformity is minimal. However, once the surgeon is experienced with hip resurfacing, it may serve as a useful strategy in the treatment of symptomatic DJD resulting from LCPD.

Summary

Hip joint arthroplasty has proven to be a very effective treatment for the symptomatic sequelae of LCPD. In cases with severe deformity or significant leg-length discrepancy, THA may be used effectively in conjunction with osteotomies, bone grafting, and modular implants.

Resurfacing hip arthroplasty is a bone-conserving procedure with inherently higher joint stability when compared with THA. However, it is a more technically challenging procedure and probably should not be used when severe proximal femoral deformity or leg-length discrepancy is present.

Authors' Disclosure Statement

The authors report no actual or potential conflict of interest in relation to this article.

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